The Case of Augustus d'Este (1794-1848): the First Account of Disseminated Sclerosis

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Introduction

THE manuscript from which this account was extracted was found amongst some papers in one of the London Sector Hospitals, and was written by Sir Augustus Frederick d'Este, a grandson of George III. He gives a vivid account of his own complaint, disseminated sclerosis, the clinical picture including transitory blindness and diplopia, absolute loss of power of locomotion followed by almost complete recovery, so that "the Chamois hunting" could be followed later, the gradual onset of paraplegia, tremors, sphincter troubles and sensory symptoms. The recital is the more remarkable, as the period covered, 1822 to 1846, is at least twenty years before any description comparable to this appeared in the medical Press. The names of many well-known doctors of the period are to be found in the account, as well as interesting details of various forms of treatment.

The manuscript book will be deposited in the library of the Royal College of Physicians, together with a few explanatory notes.

"In the month of December 1822 I travelled from Ramsgate to the Highlands of Scotland for the purpose of passing some days with a Relation for whom I had the affection of a Son. On my arrival I found him dead.—I attended his funeral:—there being many persons present I struggled violently not to weep, I was however unable to prevent myself from so doing:—Shortly after the funeral I was obliged to have my letters read to me, and their answers written for me, as my eyes were so attacked that when fixed upon minute objects indistinctness of vision was the consequence:—Until I attempted to read, or to cut my pen, I was not aware of my eyes being in the least attacked. Soon after I went to Ireland, and without any thing having been done to my eyes, they completely recovered their strength and distinctness of vision.—

In 1825 I sometimes saw imagined spots floating before my eyes. I consulted Mr. Alexander who was of opinion that I might occasionally be so troubled, but that my sight was in no danger.—In the month of January 1826 the most painful Chapter up to that period of my life occurred. I was beset by afflictions on all sides. My eyes were again attacked in the same manner as they had been in Scotland. Dr. Spangenberg sent me to Driburg, a Watering place where I drank Steel-water, bathed in it, and douched

my eyes with it: -my eyes again recovered.

In June 1827 I found my Mother in bad health at Lausanne; the heat of the country I found intolerable.—My Mother and I travelled to Venice. To my surprise I there one day found a torpor or indistinctness of feeling about the Temple of my left eye. At Florence I began to suffer from a confusion of sight:—about the 6th of November the malady increased to the extent of my seeing all objects double. Each eye had its separate vision.—Dr. Kissock supposed Bile to be the cause: I was twice blooded from the temples by leeches: -purges were administered: One Vomit, and twice I lost blood from the arm: one of the times it was with difficulty that blood was obtained.—The Malady of my eyes abated, I again saw all objects naturally in their single state. I was able to go out and walk.—Now a new disease began to shew itself: every day I found gradually (by slow degrees) my strength leaving me: I could clearly perceive each succeeding day that I went up and down the staircase with greater difficulty. When I slapped myself sharply on the loins for the time it increased my strength.—A torpor or numbness and want of sensation became apparent about the end of the Backbone and the Perineum. At length about the 4th of December my strength of legs had quite left me, and twice in one day I fell down upon the floor in attempting to go to the closed stool without assistance; I was obliged to remain on the floor until my Servant came in and picked me up. I remained in this extreme state of weakness for about 21 days, during which period I fell down about 5 times (never fainting) from my legs not being strong enough to carry my body. I never once fainted or had any sort of fit: -Debility, extreme debility was the only cause of my falling.

I now changed my Medical Advisor, putting myself under the care of a Dr. Kent, he instantly commenced a system the very opposite to the one hitherto resorted to:—Dr. Kent made me eat beef-steaks twice a day, drink London Porter and Sherry and Madeira

wines. My legs were twice a day rubbed with brushes and the torpid part of my back was rubbed twice a day (an hour each time) with a Liniment.—This new system succeeded completely. Every day I found my strength returning:—December the 14th, 15th, and 16th 1827 I was at my greatest degree of weakness; on the 25th I was able to take a short walk, and on the 21st of January (1828) I was strong enough to begin a journey from Florence (where I was during my Illness) to Rome.—On the journey I was able to walk up some steep Hills.—At Rome I continued the system recommended by Dr. Kent namely, the rubbing with flesh brushes, and the strengthening regimen. I discontinued the rubbing my back with the Liniment but my Servant used to slap me with his open hand towards the oss-sacrum.—I rode out on horseback most days, and my strength gradually returned. I never was able to run so fast as formerly, nor could I venture to dance.—

At Splugen, a small village amongst the Alps, and in its neighbourhood I remain for Six Weeks. The bracing Mountain Air of the Alps restores me so much to strength that by degrees I am enabled to take very long walks upon the mountains and to follow to a certain degree the Chamois hunting.—I could never run with my former agility and in rocky ground and in very steep places I used to complain of the feebleness of my knees. Before I left Splugen, once or twice the coming on of the torpor and want of feeling of the old place made itself slightly perceptible.—On the 3rd of September I quit Splugen: every day of my Journey I find the torpor about the Perineum and at the end of the back bone more and more perceptible: at last I get very alarmed about it—I know not what to do, whether to stop, or at Baden-Baden to try the effects of the natural Hot Baths—I resolve to go on, and reach Hannover on the —— of September. I attend the Manœuvres of a Corps of Ten Thousand men for about a fortnight. During these Manœuvres I am able to sit upon my horse for 5 and 6 hours a day but am much fatigued by it.—After the first few days I observe that my making water is attended with difficulty, particularly early in the morning, before I have had a stool, in the course of the day time the difficulty is diminished.

Immediately on my return to Hannover I consult the Surgeon Spangenberg. On the 19th of October he attempts to pass a Bougie, he finds an impediment, he withdraws the Bougie and inserts a silver Catheter with which he ruptures what he calls a slight membranous stricture.—The operation occasions me smart pain, a cold sweat breaks out on my forehead and I feel somewhat sick: some blood is the consequence but in no great quantity.—On the following day I have a Consultation between Dr. Sleiglitz and Spangenberg. They order me to take 3 times a day a decoction of herbs and flowers, and to use

every day the Shower Bath .--

I remain at Hannover from the 1st of December until the 17th always bathing every morning in the Shower Bath and drinking the tea or decoction of Flowers. During these 17 days I have observed no change in my complaint. I am able to ride for three hours at a time with pleasure, or to walk for two hours without being overfatigued: but I cannot run, or dance, and even going down stairs was unpleasant to me.

On the 17th of December I commence my journey to London:—I find that sitting for even 3 or 4 hours in a carriage increases very materially the numbness of the Perineum and extremity of my back bone, and causes a disagreeable sleepy effect all along the back part of the legs to the feet. I find that were I to attempt to travel far of a day I should be rendered incapable of proceeding.—

1829. In the beginning of August I go down to Ramsgate, and commence bathing in the Sea. I continued bathing in the Sea until the 15th of December:—during this time I take about 52 baths.—

1830. Whilst at Ramsgate I formed a liaison with a young woman—I find in my acts of connection a deficiency of a wholesome vigor. Thinking that possibly some return of Stricture (if ever there had been one) might be the cause I applied to Dr. Courtenay. He passed his largest Bougie at his first trial. There was no Stricture—he gave me some Medicine and Pills and certainly some increase of vigor was brought about.—I continued to consult him until the end of February, he orders me to be electrified by Partington, I undergo a course of Electricity.—

(Details over a period of years omitted. They deal mainly with treatment)

June 19th, 1843. What I complain of now is 1st.—that sitting produces a numbness all down the back part of my Thighs and Legs, and gives me a curious numb sensation in the lower region of the Belly.—2nd.—When standing or walking I cannot keep my balance without a Stick.

My digestion I believe to be good. I sleep well when I am not annoyed with little nervous Twitchings in my Legs or Feet, which sometimes, but seldom, is the case.—

I am naked every morning for about 30 minutes during which time I sponge myself with Cold water upon the Bidet, take Gymnastic Exercise, and sip slowly about one Pint of fresh water. My feet are rubbed and shampooed night and morning.—

I use generally the Hair-gloves. By kicking out with Strength I can exercise my legs well when steadied right and left by resting on a Chair and upon a Chest of Drawers.

I can walk accordingly as I am amused and interested by conversation:—when I am alone sometimes I walk for an hour without resting:—My walks in general are for three quarters of an hour.

December 1843.—About the 27th I saw Mr. John Scott and put it to him whether, if I were his own brother, he would not wish to talk over my Case with some one of his Medical Friends in whom he had implicit reliance. He acknowledged that such would be the case. I told him that the selection of the individual was completely with himself, in consequence of which on December the 29th he sent Dr. Farre, of No. 4 Charter House Square to visit me.—Dr. Farre read over this very book, interrogated me, and afterwards met in consultation Mr. John Scott.

January 1844.—On the 2nd of January, 1844 Dr. Farre called upon me, for the second time, and brought me the following Paper with its Prescription as the result of their Deliberation.—

London, January 1st, 1844.

'Dr. Farre met Mr. Scott in consultation on the case of Sir Augustus d'Este, and they are agreed as to its Nature, its Seat; and its Treatment, with some modifications, which Dr. Farre has suggested to make that Treatment more effectual.

'The disease is called Paraplegia and is either Active or Passive, Functional or Organic.—The transition from Functional to Organic disease is often very imperceptible; but in the Passive form of it, which is the form of the Case of Sir Augustus d'Este the duration of the functional form of the disease is often very considerable;—and Dr. Farre is of ppinion, derived from the history of the Case, that this transition is not yet Conformed.'

January 1846.—Since my return to London on the 17th of December 1845 I have regained some of my Strength which was lost at St. Leonards; but then I suffer very much indeed from sharp Spasmodic pains in my Feet and Legs:—Spasms which distress the whole System—particularly of a Morning when I get off my Bed, and at times when dressing I am obliged to stand up—supporting myself—my Sensations seem to be awful indicators that some very sad Change has taken place, or is taking place in my System. I cannot now sit for any length of time without the pressure of the hinder parts of my Thighs against the Chair occasioning Spasms.—In Bed for the last 15 months I have not enjoyed the Comfort of the recumbent Position with which almost all are blessed.—I am obliged often to change my side from a feeling of Discomfort—not amounting to Pain—but very distressing. The feeling of my Limbs in Bed are very curious; and so contrary to the Nature of Health, that I know not how to give an Idea of it.—When I stand up now for a few minutes my whole Column is affected but in a very slight degree indeed.—My sensation around my Middle is as if my Legs, having been out of joint in the Thigh sockets, had been restored to their proper place; but still were under the Influence of the Injury which they had sustained.

I must also call the attention to the fact that for the last 12 months (since about January 1845) my Hands have become slightly sufferers from my Disease; the attention being directed to them—they feel dry, and, as I should think, the Hands of a healthy person would do, after having been for some considerable time in hot water and then dried.

January 22nd, 1846.—I place myself under the Treatment of Dr. Seymour M.D. of No. 13 Charles St., Berkeley Square. He visits me—he reads this book—He expresses it as his opinion that no organic Evil exists, and that there is no reason that he can see why I should not be made well—further he says that he has known worse Cases than mine that have been materially amended.

During this month (November 1846) I suffered from attacks of Bile so far serious that they occasioned to me Sensations that led me to fear that I should fall off my Chair in some sort of Fit or Faint. With regard to my Bowels—I should say that for the last ten Weeks, they have been in a most unsatisfactory state:—during that time I have scarcely ever had Evacuations unaided by slight Aperients.

On the favourable side I have to remark that the Sensitiveness of the Skin around my Hips and down my Legs is decidedly finer; for I cannot bear the friction of the Horsehair Gloves as I used to do. The substance of the foregoing Statement I sent to Dr. Seymour by Letter on or about the 20th (of November) and on the 25 I received the Answer.—In Dr. Seymour's Letter there is as follows:—'On the whole I think you had better leave off the Cantharides until We meet; but you will then have to resume it—as I feel very confident by Perseverance of an ultimate Recovery.'

(The last entry 1846.)

Monday December 17th. Having received a Present of Indian Moccassins I put them on:—and I walk in them without my Left Foot, which, some time ago always turned over outwards at the Ankle joint unless supported by a Steel Up right, showing any disposition so to do. Surely this is a decided Improvement. Thanks to The Almighty!"

Sir Augustus d'Este died early in 1848, in his 54th year. The cause of his death is unknown, but it is clear from his last diaries that his spasticity became so marked that he was unable to walk up any stairs and spent most of his time in his "chair on wheels." His spasms frequently caused broken nights, but they were not the only reason of his insomnia, for one entry reads: "During the past Night, I suffer from Bugs and Spasms. I get up sadly late".